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AMERICAN OTOLOGICAL SOCIETY MEETING PAPERS

FIBROUS DYSPLASIA OF THE TEMPORAL BONE: TEN NEW CASES DEMONSTRATING THE SPECTRUM OF OTOLOGIC SEQUELAE

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ABSTRACT

Fibrous dysplasia involving the temporal bone is unusual. The most common initial findings are stenosis of the external auditory canal and conductive hearing loss. The frequency of sensorineural hearing loss and facial paresis as complications of fibrous dysplasia have been poorly documented in the past. Forty-three cases of fibrous dysplasia of the temporal bone previously published in the otolaryngologic literature are reviewed, and 10 new cases are reported. In addition to age, sex, and frequency of various presenting complaints, the audiometric, radiographic, and surgical data are evaluated in both groups to better describe the manifestations of disease, degree and type of hearing loss, and success of surgical intervention. In nearly 70% of cases in this study, fibrous dysplasia was monostotic. Although most patients had a conductive hearing loss, 17% of patients demonstrated profound sensorineural hearing loss ascribable to the lesion, and facial nerve sequelae were noted in nearly 10% of cases. Cholesteatoma complicated almost 40% of cases, usually in the form of a canal cholesteatoma. Ten new cases of temporal bone fibrous dysplasia are described not only to further clarify the spectrum of otologic sequelae but also to help illustrate available treatment options. In addition, this report documents, in three new cases, the previously undescribed progression of conductive hearing loss to profound sensorineural deafness secondary to fibrous dysplasia.

Fibrous dysplasia is characterized by a slow, progressive replacement of a localized area of bone by an abnormal proliferation of isomorphic fibrous tissue intermixed with poorly formed, haphazardly arranged trabeculae of woven bone often described histopathologically as demonstrating a "Chinese letter" or "jigsaw puzzle" configuration. The name fibrous dysplasia was first suggested by Lichtenstein in 1938, in an attempt to distinguish the fibro-osseous disease from other bony lesions, such as giant cell tumor, ossifying fibroma, fibrosarcoma, osteodystrophy, osteoid osteoma, fibroma, and "osteitis fibrosa cystica" resulting from hyperparathyroidism and disordered calcium metabolism.[1]

The lesions of fibrous dysplasia appear in three distinctive clinical patterns: involvement of a single bone (monostotic form), multiple bones (polyostotic form), and that occurring in the polyostotic form with pigmentation and endocrinologic abnormalities (Albright syndrome). In all clinical patterns, the bony lesions cause localized defects in cortical and cancellous bone ranging from a few centimeters in size to massive lesions that distort the normal contour of the involved bone.[2] Monostotic fibrous dysplasia accounts for 70 to 75% of all cases and, in order of frequency, affects the ribs, femur, tibia, maxilla, mandible, calvarium, humerus, and other bones. Although the lesions are often asymptomatic, relatively small lesions of the face or skull may cause disfigurement.[3] Polyostotic fibrous dysplasia accounts for about 30% of cases and tends to occur at an earlier age than the monostotic form. Lesions are usually limited to a single limb or one side of the skeleton. In milder cases of the polyostotic form, craniofacial

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lesions occur in about 50% of patients, whereas in more severe cases involvement of the face and skull occurs in nearly 100% of cases.[4] In 1937, Albright described the relatively rare variant, occurring in approximately 3% of cases, characterized by the triad of polyostotic fibrous dysplasia, pathologic skin pigmentation, and precocious puberty in females.[5] However, other endocrine disorders complicate the skeletal lesions of Albright syndrome, including hyperthyroidism, Cushing syndrome, hyperparathyroidism, and pituitary-induced abnormal skeletal growth.[6]

The etiology of fibrous dysplasia is controversial. Putative causes include dysplasia caused by abnormal enzymatic activity in bone-forming mesenchyme,[7] disordered metabolism of calcium and phosphorus,[8] and hyperplasia of osteoblasts.[9] Schlumberger hypothesized that the monostotic form of the disorder may be unrelated to both Albright syndrome and polyostotic fibrous dysplasia and instead is caused by a disturbance of the normal reparative processes following bone injury.[10] The organs most often affected in Albright syndrome are under the control of trophic factors that modulate the intracellular concentration of cyclic adenosine monophosphate (cAMP). Recent evidence has suggested that in Albright syndrome there is autonomous hyperfunction of these affected glands and organs due to abnormal intracellular regulation of cAMP or protein kinase A.[11]

The most common clinical manifestations of fibrous dysplasia of the temporal bone are progressive stenosis of the external auditory canal and conductive hearing loss.[12] Radiographic study of these lesions usually demonstrates a homogeneous radiodensity involving any combination of the tympanic, mastoid, squamous, and petrous portions of the temporal bone. Three radiographic patterns have been described in fibrous dysplasia of the skull and facial bones by Fries: pagetoid (56%), a mixture of dense and radiolucent areas of fibrosis; sclerotic (23%), homogeneously dense; and cystic (21%), a spherical or ovoid lucency surrounded by a dense boundary.[13] A predominance of osseous elements renders the lesions more opaque radiographically. An abundance of fibrous elements produces a radiolucent or cystic appearance. A mixture of fibrous and bony elements produces the classic "ground-glass" appearance.[14] The differential diagnoses of these lesions seen in radiographs of the temporal bone include non-ossifying fibroma, unicameral cyst, Paget disease of bone, osteochondroma, giant cell granuloma, exostosis, osteoma, aneurysmal bone cyst, ossifying fibroma, as well as primary or metastatic sarcomatous tumors. The definitive diagnosis of fibrous dysplasia of the temporal bone is made by correlation of radiographic and pathologic findings. Although fibrous dysplasia may appear clinically most similar to ossifying fibroma, it can be distinguished microscopically by a woven pattern of bone formation rather than the lamellar pattern typical of the more infrequent ossifying fibroma.[15]

The treatment of fibrous dysplasia of the temporal bone has traditionally been conservative and directed toward the management of stenosis of the external auditory canal. The clinical course of fibrous dysplasia is unpredictable. Some lesions remain stable for many years, whereas others can progress rapidly. However, fibrous dysplasia usually demonstrates a chronic, slowly progressive growth pattern and tends to show less activity in older patients. Spontaneous neoplastic transformation into sarcoma has been described in 0.4% of cases.[16] Radiation therapy is contraindicated in the management of fibrous dysplasia because it may increase the incidence of malignant degeneration.[17]

The first report in the English literature of monostotic fibrous dysplasia involving the temporal bone was provided by Schlumberger in 1946.[10] In this review of 67 cases, he found only one in which the temporal bone was involved. In this case, a 12-year-old boy presented with frequent aural discharge, otalgia, and a retroauricular mass. Exploration of the ear revealed an extremely vascular mastoid bone, and the contents of the bony lesion were gritty with the consistency of dessicated cheese. In 1952, Brunner presented the earliest reports of polyostotic fibrous dysplasia affecting the temporal bone, in two patients with stenosis of the ear canal, hearing loss, and significant facial deformity.[18] The first documentation of temporal bone involvement in a case of Albright syndrome was by Barrionuevo et al in 1980.[19] They described a 3-year-old girl with a 2-year history of breast hypertrophy, vaginal bleeding, a large abdominal and pelvic café au lait spot, increased urinary gonadotropin levels, and radiographic evidence of polyostotic fibrous dysplasia involving the left mastoid bone.

Ten previously unreported cases of fibrous dysplasia of the temporal bone are presented in this report bringing the total number of cases documented in the English otolaryngologic literature to 53. In an attempt to better characterize the clinical manifestations and otologic sequelae of fibrous dysplasia of the temporal bone, both new and historical cases were reviewed.

METHODS

A review of the English literature was performed to identify cases of fibrous dysplasia of the temporal bone that included specific information regarding: (1) age at which symptoms were first noted, (2) gender of patient, (3) nature of presenting symptoms, (4) otologic examination, (5) nature of hearing loss, (6) form of disease (i.e., monostotic, polyostotic, Albright syndrome), (7) history of ear surgery, (8) presence of cholesteatoma, and (9) status of the facial nerve.

In an attempt to identify previously unreported cases of fibrous dysplasia of the temporal bone, the files of the departments of medical records, pathol-

ogy, and radiology at the Massachusetts Eye and Ear Infirmary and the University of Vermont Medical Center were searched. Similar pertinent historical information was gathered by retrospective chart review of known cases at both institutions.

RESULTS

Historical review was limited to previously reported cases in which there was adequate documentation of otologic symptoms and findings. This yielded 43 cases of fibrous dysplasia involving the temporal bone previously reported in 32 publications (Table 1).[18–49] A summary of these 43 cases is presented in Table 2.

The form of disease expression, as documented by radiographic, serologic, and physical examination, included monostotic fibrous dysplasia in 69.8% of cases, polyostotic fibrous dysplasia in 23.2% of cases, and Albright syndrome in three cases (7%). Only one of the cases classified as Albright syndrome demonstrated the classic triad of polyostotic fibrous dysplasia, hyperpigmentation, and precocious puberty in a female. Two other cases categorized as Albright syndrome lacked precocious puberty and the female gender, but displayed the well-known association of polyostotic fibrous dysplasia, hyperpigmentation, and endocrine disorders (hyperthyroidism). The mean age of presentation of fibrous dysplasia of the temporal bone was 19.6 years (range, 9 mo–54 yr), and 60.4% of patients were male.

The presenting symptoms included hearing loss in 79.1%, temporal bone mass in 25.6%, unilateral otorrhea in 13.9%, otalgia in 6.9%, and trismus in 2.3%. Otologic findings included stenosis of the external auditory canal, and in many cases this was misdiagnosed as exostosis or osteoma. A conductive hearing loss was present in 79% of cases, sensorineural hearing loss in 14%, and normal hearing in 7%. Progression from conductive hearing loss to sensorineural hearing loss has not been previously reported. Of the six previously reported cases of sensorineural loss (14% of total), four were associated with monostotic fibrous dysplasia, and two with the polyostotic form. Imaging studies (polytomography and computed tomography [CT]) of the six cases of sensorineural hearing loss demonstrated encroachment of the internal auditory canal in five cases and erosion of the lateral semicircular canal in one case.[29]

Eighty-six percent of previously reported cases underwent at least one surgical procedure during their course. Restenosis of the external auditory canal occurred in 48.6% of cases. The incidence of restenosis of the canal was 88.9% in patients who underwent transcanal or endaural canaloplasty without mastoidectomy and 0% in cases treated with postauricular canaloplasty with mastoidectomy that included at least 6 months follow-up information. Cholesteatoma complicated 39.5% of cases with canal stenosis, and 82.4% of such cholesteatomas were limited to the external auditory canal and lateral to an intact drum. Facial nerve paresis or paralysis occurred in 9.3% of cases (4 patients). Two patients had temporary or partial paresis secondary to concomitant chronic otitis media. Two other patients demonstrated complete and permanent facial paralysis secondary to encroachment of the facial canal by fibrous dysplasia.

CASE REPORTS

Clinical data from 10 new cases of fibrous dysplasia of the temporal bone are presented in Table 3 and summarized in Table 4.

Case 1

A bony right postauricular mass and hearing loss was identified by this 4-year-old boy's pediatrician. An audiogram at the time demonstrated an ipsilateral conductive hearing loss of 40 dB. Polytomography and CT scanning demonstrated a fibro-osseous lesion of the inferior and anterior bony external canal that extended to the mastoid tip, consistent with monostotic fibrous dysplasia (Fig. 1). No further medical intervention occurred until age 12, at which time this boy initially presented to the Emergency Department of the Massachusetts Eye and Ear Infirmary with right otalgia and otorrhea. There had been previous recurrent episodes of external otitis. Physi-

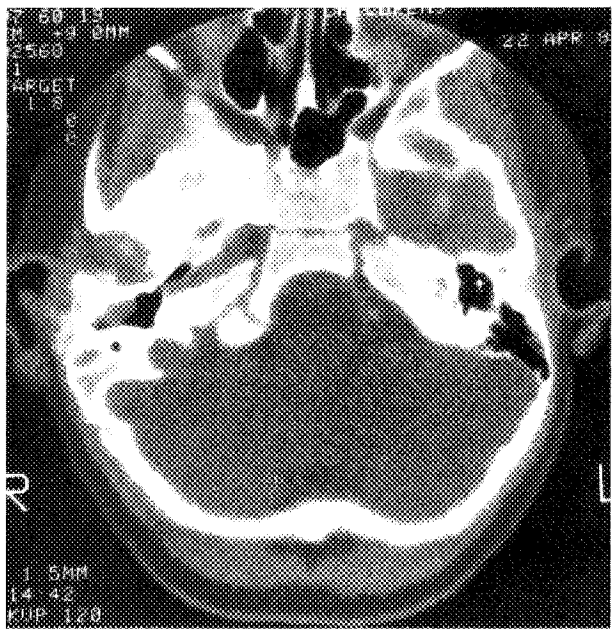


Figure 1. Axial CT scan (case 1) demonstrating right external auditory canal stenosis secondary to fibrous dysplasia.

Table 1. Previously Published Case Reports of Temporal Bone Fibrous Dysplasia

Author	Year	Age (Yr)	Sex	Symptoms	Ear Examination	Hearing	Previous Surgery	Disease	Cholesteatoma	Facial Nerve	Author's Surgery	Follow-Up
Reddy et al[49]	1994	5	F	Vertigo	Normal	SNHL	None	Monostotic	None	Paralysis	None	N/A
Kessler et al[48]	1990	15	M	HL	Stenosis	Cond	CP	Monostotic	None	NL	None	Re-stenosed
Talmi et al[47]	1989	15	F	Otitis	Stenosis	Cond	None	Monostotic	Canal	NL	CP	Not known
Pouwels et al[46]	1988	18	F	HL, otitis	Stenosis	Cond	CP × 2	Polyostotic	Mastoid	NL	PAM CP	NED 2 yr
Younus et al[45]	1987	9 mo	M	Mass	Stenosis	Cond	None	Monostotic	None	NL	PAM CP	Not known
Smouha et al[44]	1987	54	M	HL	Stenosis	Cond	CP	Monostotic	None	NL	PAM CP	NED 3 yr
"	1987	14	M	HL	Stenosis	Cond	Biopsy	Monostotic	Canal	NL	PAM CP	NED 2 yr
"	1987	47	F	HL	Stenosis	Cond	None	Monostotic	Canal	NL	PAM CP	NED 2 yr
Sataloff et al[43]	1985	9	F	HL	Stenosis	Cond	None	Polyostotic	None	NL	CP, PAM CP	Not known
Wolfson et al[42]	1984	26	F	HL	Stenosis	Cond	None	Polyostotic	Canal	NL	PAM CP	Not known
Lambert et al[41]	1984	7	F	HL	Stenosis	Cond	CP	Monostotic	None	NL	None	Re-stenosed
Nager et al[40]	1984	3	M	Otitis	Stenosis	SNHL	Mastoid × 4	Polyostotic	Canal	NL	PAM CP	Not known
"	1984	7	M	HL, otitis	Stenosis	Cond	Mastoid	Monostotic	Canal	NL	PAM CP	Not known
Schrimpf et al[39]	1982	32	M	HL	Stenosis	Cond	None	Monostotic	Canal	NL	PAM CP	NED 6 mo
Nager et al[38]	1982	10	M	HL	Stenosis	Cond	CP	Albright	Canal	NL	CP	Not known
"	1982	39	M	HL	Stenosis	Cond	None	Albright	Canal	NL	None	N/A
"	1982	10	M	HL	Stenosis	Cond	Biopsy	Monostotic	None	NL	PE tube	Not known
Levine et al[37]	1981	27	F	HL, otitis	Stenosis	Cond	CP	Polyostotic	None	NL	CP × 2, TBR	NED 1 yr
Barrionuevo et al[19]	1980	11	F	Mass	Normal	Normal	None	Monostotic	None	NL	None	N/A
"	1980	19	M	Otitis	Stenosis	Cond	Mastoid × 2	Monostotic	Mastoid	Palsy	PAM CP, drain abscess	NED 1 yr
"	1980	3	F	Precocious puberty	Normal	Normal	None	Albright	None	NL	None	N/A
Ward et al[36]	1975	17	F	HL	Stenosis	Cond	Mastoid	Monostotic	None	NL	CP, PAM CP	NED 2 yr
Williams et al[35]	1975	10	F	HL	Stenosis	Cond	None	Monostotic	None	NL	PAM CP	NED 10 yr
Talbot et al[34]	1974	12	M	HL	Stenosis	Cond	None	Monostotic	None	NL	CP × 2	Re-stenosed
Chatterji et al[33]	1974	30	F	HL, mass	Stenosis	SNHL	None	Monostotic	None	NL	None	N/A
Stecker et al[32]	1971	11	M	HL	Normal	Cond	None	Monostotic	None	NL	PAM CP	NED 6 mo
Sharp et al[31]	1970	11	M	HL, mass	Stenosis	Cond	Mastoid	Monostotic	Canal	Palsy	Mastoid × 3	Not known
"	1970	30	M	HL, otalgia	Stenosis	Cond	PAM CP	Monostotic	Canal	NL	PAM CP	Not known
Tembe[30]	1970	18	M	HL	Stenosis	Cond	None	Polyostotic	Canal	NL	CP	NED 14 mo
Cohen et al[29]	1969	8	F	Mass	Stenosis	SNHL	Mastoid	Polyostotic	None	Paralysis	Mastoid × 2	NED 12 yr
"	1969	9	M	Mass	Stenosis	Cond	PAM CP	Monostotic	None	NL	PAM CP	NED 1 yr
Shiffman et al[28]	1967	29	M	HL, trismus	Stenosis	Cond	None	Monostotic	Canal	NL	PAM CP	NED 8 mo
Samy et al[27]	1967	43	M	HL, mass	Stenosis	SNHL	None	Monostotic	None	NL	PAM CP	Not known
"	1967	9	M	HL	Stenosis	Cond	CP	Polyostotic	None	NL	CP	Not known
Basek[26]	1967	37	M	HL	None	Cond	None	Monostotic	None	NL	CP	NED 7 mo
Fluur et al[25]	1966	16	M	Mass	None	None	None	Monostotic	None	NL	Mastoid	Not known
Wong et al[24]	1965	13	F	HL, mass	Stenosis	Cond	Biopsy	Monostotic	Canal	NL	CP	NED 9 mo
Sussman[23]	1961	16	F	HL, otalgia	Stenosis	Cond	Biopsy	Monostotic	None	NL	None	N/A
Kearney[22]	1959	21	M	HL, mass	Stenosis	Cond	CP	Monostotic	Mastoid	NL	PAM CP	NED 3 yr
Skolnik et al[21]	1958	54	F	HL	None	SNHL	None	Monostotic	None	NL	None	N/A
Brunner[18]	1952	27	M	HL	Stenosis	Cond	None	Polyostotic	None	NL	CP	Re-stenosed
"	1952	35	M	HL	Stenosis	Cond	None	Polyostotic	None	NL	None	N/A
Towson[20]	1950	14	M	HL	Stenosis	Cond	Biopsy	Monostotic	None	NL	Mastoid	Death

HL = hearing loss, SNHL = sensorineural hearing loss, Cond = conductive, CP = canaloplasty, PAM = postauricular mastoidectomy, NED = no evidence of disease, TBR = temporal bone resection

Table 2. Clinical Summary of Historical Cases

Age (mean)	19.6 years
Sex	60.4% male
Chief complaint	79.1% hearing loss 25.6% mass 13.9% draining ear 6.9% otalgia 2.3% trismus
Otologic examination	83.7% canal stenosis
Hearing	79.1% conductive loss 13.9% sensorineural loss 7.0% normal hearing
Fibrous dysplasia	69.8% monostotic 23.2% polyostotic 7.0% Albright syndrome
Cases having surgery	86% at least one procedure
Surgical failure	48.6% re-stenosis
Canaloplasty failures	88.9% re-stenosis
Cholesteatoma	39.5% of cases
Canal cholesteatoma	82.4% of cholesteatomas
Facial nerve	4.6% paresis 4.6% complete paralysis

cal examination revealed purulent otorrhea and marked stenosis of the bony external canal. The Weber test lateralized to the involved ear. The remainder of the head and neck examination was within normal limits. His external otitis was successfully managed medically, and plans were made for surgical repair of his stenotic ear canal, including postauricular mastoidectomy, canaloplasty, and split-thickness skin grafting. The patient, however, was lost to follow-up.

Case 2

This 9-year-old female initially presented to the Massachusetts Eye and Ear Infirmary after failing a school audiogram. The patient reported right sided hearing loss, occasional otalgia, and frequent ipsilateral swimmer's ear. She denied vertigo or tinnitus. The right external auditory canal was nearly totally obstructed by a bony mass that emanated from the floor of the canal and was covered with normal skin. The remainder of her head and neck examination was normal. An audiogram revealed a conductive hearing loss of 60 dB on the right. Computed tomography and magnetic resonance imaging (MRI) demonstrated a cystic mass that was centered in the hypotympanum, bulged into the lateral wall of the jugular foramen, and involved the middle ear and mastoid air cells (Figs. 2 and 3). Biopsy of the canal mass was performed via a postauricular approach. The final pathologic diagnosis was consistent with cystic monostotic fibrous dysplasia (Fig. 4). This patient was managed expectantly and followed with serial CT scans. Six months later, an audiogram demonstrated a profound sensorineural hearing loss on the right side (Fig. 5). A follow-up CT scan demonstrated probable invasion of the vestibule by the bony mass. Because of the progressive nature of this tumor, a decompression of the jugular foramen and internal auditory canal via a modified infratemporal fossa approach was performed due to signs of progressive lower cranial nerve compression.

Case 3

This 33-year-old female was hospitalized with severe, intermittent abdominal pain due to acute intermittent porphyria. Her medical evaluation included MRI of the head, which demonstrated a mass in the

Table 3. Ten New Cases of Temporal Bone Fibrous Dysplasia

Case Number	*Age (Yr)*	*Sex*	*Symptoms*	*Ear Examination*	*Hearing*	*Previous Surgery*	*Disease*	*Cholesteatoma*	*Facial Nerve*	*Surgery*	*Follow-Up*
1	4	M	HL, mass	Stenosis	Cond	None	Monostotic	None	NL	None	Lost
2	9	F	HL	Stenosis	Cond→SNHL	Biopsy	Monostotic	None	NL	Planned	Pending
3	33	F	Mass	Normal	Cond	None	Monostotic	None	ENoG 70%	None	Pending
4	12	F	HL	Stenosis	Cond	CP × 3	Polyostotic	Canal	NL	PAM CP	NED 9 mo
5	12	F	HL, trismus	Stenosis	Cond	CP × 4	Monostotic	None	NL	TMJ	NED 3 mo
6	5	F	HL	Stenosis	Cond	None	Polyostotic	None	NL	Biopsy	Pending
7	56	F	Mass	Stenosis	Cond	None	Monostotic	None	NL	None	Lost
8	10	F	HL	Stenosis	Cond	None	Polyostotic	None	NL	Maxilla	Lost
9	26	F	HL	Stenosis	Cond→SNHL	CP × 3	Monostotic	Canal	NL	CP	Stable 5 yr
10	2	M	Otitis	Stenosis	Cond→SNHL	PAM CP	Monostotic	None	Paralysis	VII decomp	Stable 1 yr

HL = hearing loss, SNHL = sensorineural hearing loss, CP = canaloplasty, PAM = postauricular mastoidectomy, NED = no evidence of disease

Table 4. Clinical Summary of New Cases

Age (mean)	16.9 years
Sex	80% female
Chief complaint	80% hearing loss 50% mass 30% draining ear 10% otalgia 10% trismus
Otologic examination	80% canal stenosis
Hearing	60% conductive loss 30% conductive → SNHL 10% normal hearing
Fibrous dysplasia	70% monostotic 30% polyostotic 0.0% Albright syndrome
Previous canaloplasty	30%
Surgical failure	100% re-stenosis
Cholesteatoma	20% of cases
Canal cholesteatoma	100% of cholesteatomas
Facial nerve	10% abnormal ENoG, normal examination 10% complete paralysis
VII decompression	Grade III recovery (one case)

left temporal area, and CT, which demonstrated a mass filling the bony confines of the temporal bone with the classic ground-glass appearance of fibrous dysplasia (Fig. 6). A bone scan demonstrated that the temporal bone lesion was an isolated abnormality, consistent with a diagnosis of monostotic fibrous dysplasia. The patient was referred to the Massachusetts Eye and Ear Infirmary for further care. At her initial clinic visit she denied hearing loss, tinnitus, vertigo, or weakness of the facial nerve. Physical examination revealed a bony mass in the left postauricular and suboccipital areas. Her ear canals, tympanic membranes, and audiogram were normal, bilaterally. The remainder of her head and neck examination was normal. Electroneurography (ENoG) demonstrated a compound motor action potential on the involved side to be 70% of that on the normal left side. At this time the patient is being managed expectantly with plans for decompression of the facial nerve if future examinations reveal continued facial nerve degeneration.

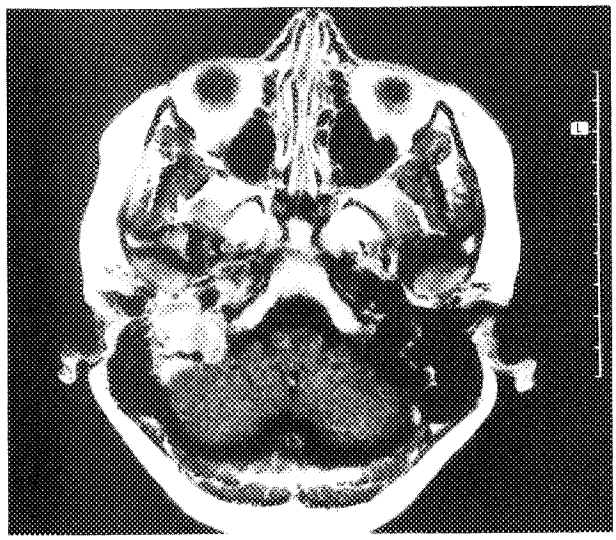


Figure 3. T1-weighted, post gadolinium MRI scan (case 2) of cystic fibrous dysplasia of right hypotympanum and jugular fossa region with extension to posterior fossa.

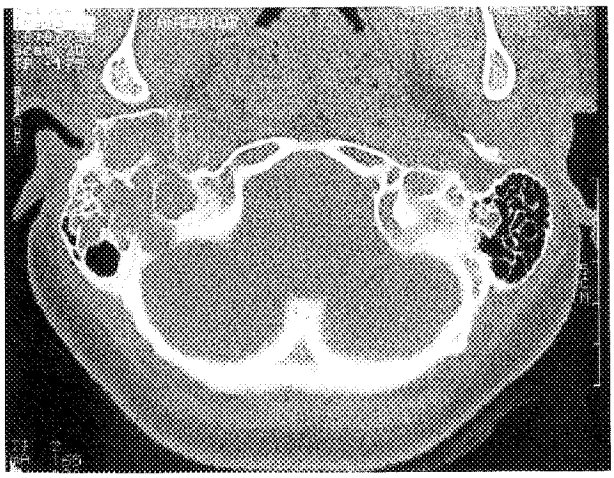


Figure 2. Axial CT scan (case 2) with obliteration of right external auditory canal, mastoid, and hypotympanic region by cystic fibrous dysplasia.

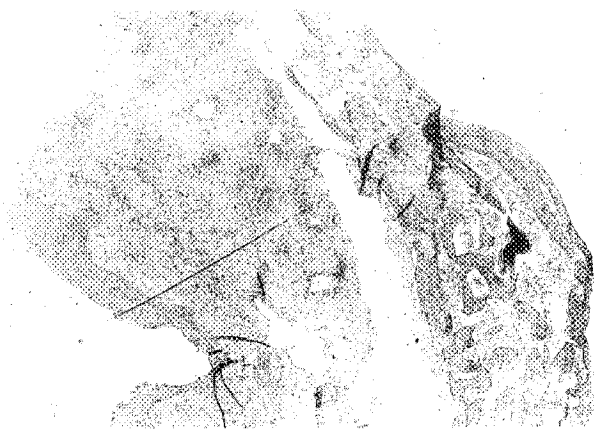


Figure 4. Surgical biopsy (case 2) of cystic fibrous dysplasia. Note rim of dysplastic bone surrounding a large central region containing predominantly fibrous tissue and inflammatory cells.

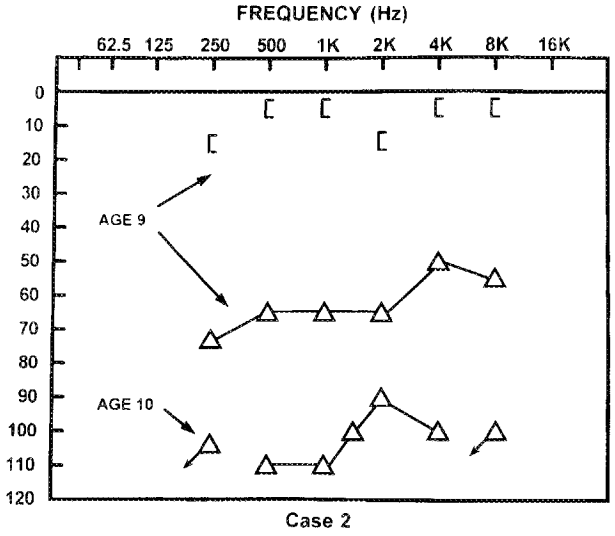


Figure 5. Composite audiogram (case 2) demonstrating progression from conductive hearing loss to profound sensorineural hearing loss over 1 year.

Case 4

At the age of 12, this female patient was diagnosed with polyostotic fibrous dysplasia involving the right temporal bone and mandible. Three canaloplasty procedures at ages 12, 19, and 43 years, for stenosis and conductive hearing loss failed because of rapid restenosis. At age 44, this patient was referred to Massachusetts Eye and Ear Infirmary with recurrent hearing loss and right aural pressure and fullness. Facial asymmetry due to marked fullness in the region of the right cheek and temporal bone was found. There was complete stenosis of the right ear canal. The remainder of her head and neck examination was normal. An audiogram demonstrated a conductive hearing loss of 60 dB on the right. A CT scan demonstrated complete bony stenosis of the external auditory canal. A right postauricular mastoidectomy was performed. The malleus and incus were fixed by the lesion, and a cholesteatoma was present in the attic. A canal wall down type-III tympanoplasty, anterior canaloplasty, and split-thickness skin grafting were performed. Pathology was consistent with fibrous dysplasia. Follow-up at 9 months showed a widely patent external auditory canal.

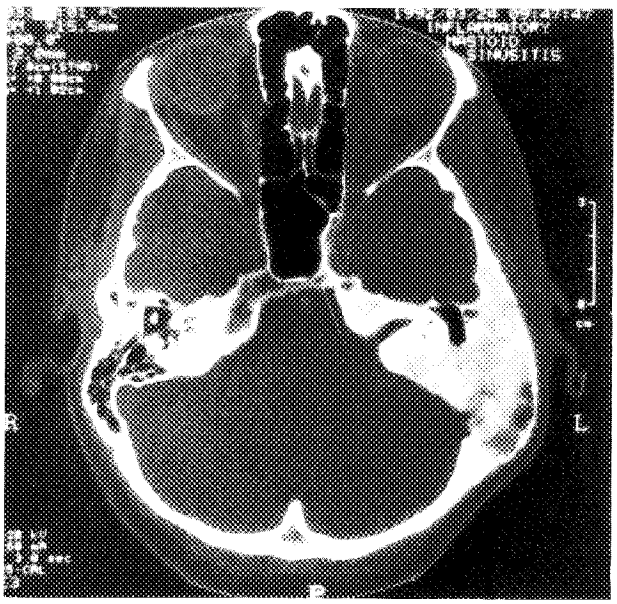


Figure 6. Axial CT scan (case 3) of Pagetoid fibrous dysplasia replacing confines of left temporal bone with preservation of otic capsule structures. Note ground-glass appearance of lesion secondary to a mixture of fibrous and bony elements.

Case 5

At the age of 12, this female patient was diagnosed with monostotic fibrous dysplasia of the right temporal bone, causing hearing loss, otalgia, and otorrhea. Over the next 28 years she underwent four canaloplasty procedures. Shortly after her last operation, she developed marked trismus and was referred to the oral surgery service at the Massachusetts General Hospital. On physical examination, she had a patent external auditory canal and decreased hearing on the right side. In addition, she had marked trismus with a maximal inter-incisor distance of 1 cm. The remainder of her head and neck examination was normal. Polytomography of the right temporal bone demonstrated fibrous dysplasia involving the bony external canal, mastoid, tympanic, and squamosal portions of the temporal bone. There was narrowing of the temporomandibular joint (TMJ) space anteriorly secondary to fibrous dysplasia. The patient underwent a successful arthroplasty of the right TMJ, with bony recontouring of the zygomatic and temporal bones, and coronoidectomy. Postoperatively, she achieved relief from trismus and now maintains a near normal range of motion of her jaw with physiotherapy.

Case 6

This female patient developed stenosis of the right external auditory canal at age 5 years. A mild hearing loss developed during her adult life. At age 42, she noticed pain and swelling in the right temporal and supraorbital regions. At age 44, polytomography demonstrated several lytic lesions with a ground-glass appearance centered at the frontal, zygomatic, and lateral temporal bone area, with destruction of the bony external auditory canal. She was referred to the Massachusetts Eye and Ear Infirmary with a tentative diagnosis of polyostotic fibrous dysplasia. Physical examination revealed diffuse swelling above and behind the right ear with occlusion of the canal. Audiometry revealed an ipsilateral conductive loss

with an SRT of 60 dB. A CT scan demonstrated involvement of the frontal, temporal, and parietal bones with no intracranial extension. A postauricular biopsy was performed that demonstrated small narrow spicules of woven bone surrounded by a fibrous matrix consistent with fibrous dysplasia. The patient was advised that it was not possible to completely remove the lesion and that therapy should be limited to correction of either cosmetic or functional defects. She did not pursue corrective canaloplasty. Aside from her hearing loss, she has been otherwise asymptomatic.

Case 7

This 56-year-old female initially presented to the Massachusetts Eye and Ear Infirmary with a 4-month history of a nontender mass over the right temporal-zygomatic region. Physical examination revealed a hard bony mass and narrowing of the external auditory canal. The remainder of her head and neck examination was within normal limits. Audiometry demonstrated a conductive hearing loss. A CT scan showed significant thickening of the floor of the middle cranial fossa and temporal bone from the parasellar region and petrous apex to the squamous bone (Fig. 7). A radiographic diagnosis of monostotic fibrous dysplasia was then made, and auditory brainstem response (ABR) testing confirmed the absence of retrocochlear involvement of the auditory nerve. Blink reflex latencies and ipsilateral-contralateral reflex-latency differences did not exceed normal tolerance limits, suggesting no involvement of the facial nerve. The patient has been lost to follow-up.

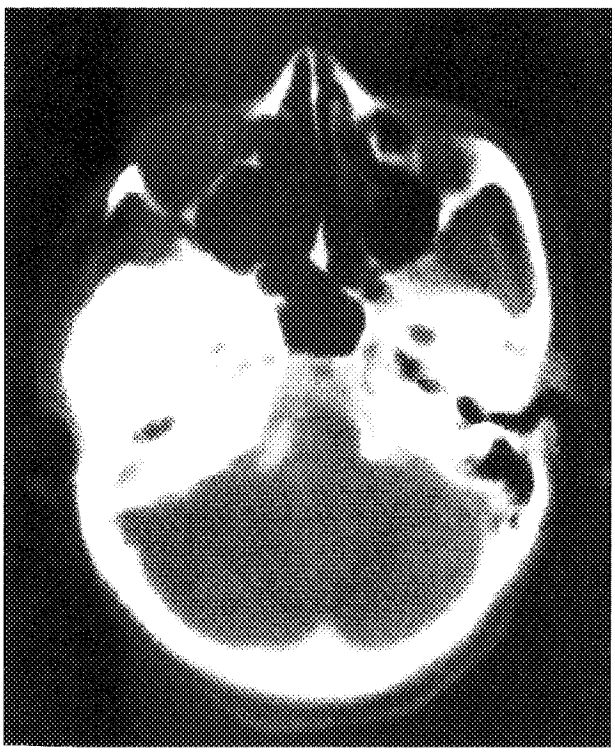


Figure 7. Early generation axial CT scan (case 7) that reveals sclerotic fibrous dysplasia that has led to thickening of the floor of the right middle cranial fossa and temporal bone from the parasellar and petrous apex region to the external auditory canal.

Case 8

At the age of 10 years, this patient was referred to the Massachusetts Eye and Ear Infirmary by her school for evaluation of facial asymmetry. Her past medical history was negative for trauma and otherwise unremarkable. Physical examination revealed marked protrusion of the right frontal region causing the orbit to be pushed down and forward 1 cm. Polytomography demonstrated an intense sclerotic reaction involving the right frontal bone and greater and lesser wings of the sphenoid, with slight narrowing of the right optic foramen. The radiographic appearance was suggestive of fibrous dysplasia. Ophthalmology examination demonstrated normal optic disks and maculae. An evaluation by Dr. Albright's endocrine service at Massachusetts General Hospital failed to demonstrate abnormal skin pigmentation, signs of endocrine dysfunction, or precocious puberty. A diagnosis of polyostotic fibrous dysplasia was made. The patient was fitted with corrective glasses and followed closely. At age 38, the patient was seen in the otology clinic complaining of decreased hearing and right aural fullness. The physical examination revealed canal stenosis and external otitis on the right. Polytomography demonstrated a large area of fibrous dysplasia involving the right parietal and frontal bones and part of the left frontal bone. The frontal sinus was obliterated by the lesion. The right external canal, tympanic cavity, and epitympanic recess were also involved. She was treated with antibiotics and decongestants and noted relief from her symptoms. Due to progressive right maxillary asymmetry she underwent surgical reduction of her maxilla via a sublabial approach at age 39 and a hemimaxillectomy at age 42. Soon thereafter, she was lost to follow-up.

Case 9

This female patient first noted left-sided hearing loss at age 26. Physical examination at an outside institution revealed canal stenosis. She then underwent five transcanal and endaural canaloplasty procedures over the next 7 years, each of which resulted in rapid re-stenosis. At age 31, she was referred to the Massachusetts Eye and Ear Infirmary with a diagnosis of monostotic fibrous dysplasia of the temporal bone. Polytomography demonstrated bony changes consistent with fibrous dysplasia involving the entire temporal bone, including the external auditory canal. An audiogram demonstrated a 50-dB conductive hear-

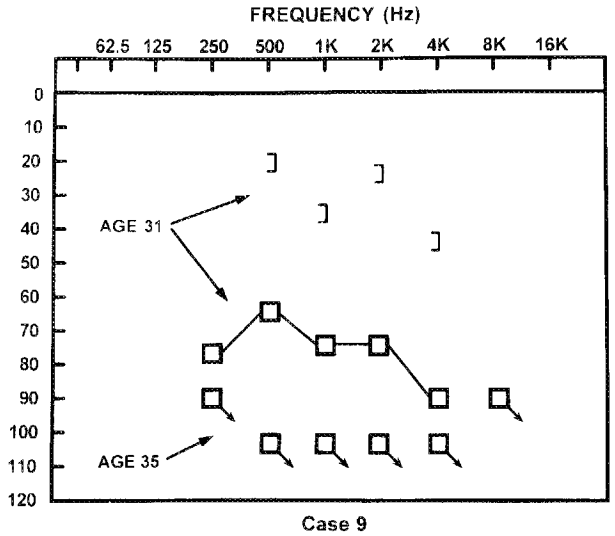


Figure 8. Composite audiogram (case 9) from a patient with long-standing fibrous dysplasia of the left temporal bone who developed acute vertigo and profound sensorineural hearing loss in the involved ear. Over the preceding 9 years, the patient had demonstrated a stable 50-dB conductive hearing loss.

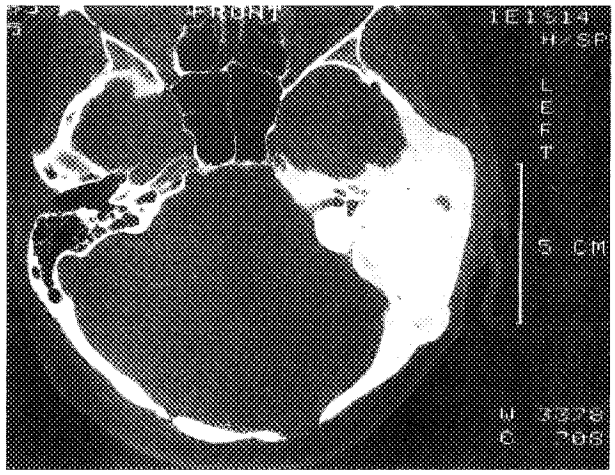


Figure 10. Axial CT scan (case 9) with extensive replacement of normal left temporal bone architecture by a component of predominantly sclerotic fibrous dysplasia. Note encroachment of the vestibule and narrowing of the internal auditory canal in this patient with sensorineural hearing loss. This lesion produced a marked external deformity and required surgical curettage for cosmesis.

ing loss. At age 35, this patient experienced an episode of severe vertigo that lasted several days. Follow-up audiometric evaluation demonstrated a new profound sensorineural hearing loss in the involved ear (Fig. 8). Over the next 10 years, yearly radiographic follow-up demonstrated minimal progression of her disease. At age 46, a bony protuberance from the left external auditory canal was removed for cosmetic purposes, and pathology was consistent with fibrous dysplasia (Fig. 9). A CT scan obtained at the time showed diffuse involvement of the temporal bone with a mixed lytic and sclerotic process (Fig. 10). Although the facial canal was obscured by the sclerotic process, facial nerve function has remained normal. Over the ensuing 5 years, the patient has remained well and without discomfort, as the growth appears to have stabilized.

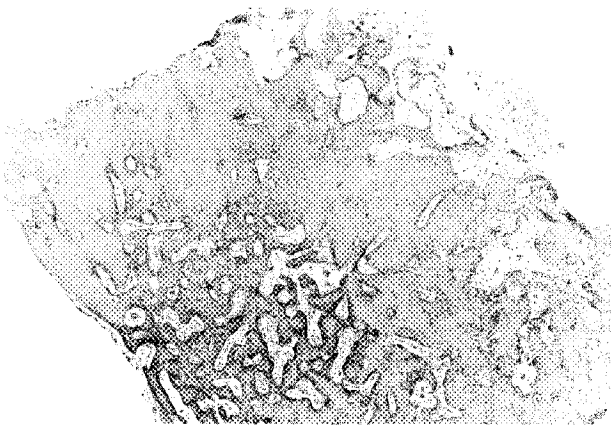


Figure 9. Surgical biopsy (case 9) from patient with monostotic fibrous dysplasia of the left temporal bone. Note dense proliferation of predominantly woven bone with minimal fibrous elements; consistent with the opaque radiographic pattern of the sclerotic variety of fibrous dysplasia (see Fig. 10).

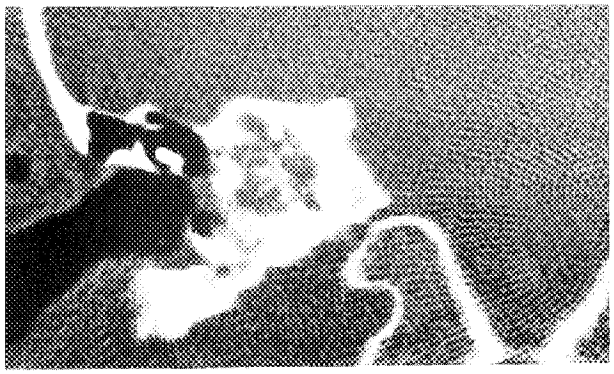
Case 10

At age 2 years, this male patient was referred to the otolaryngology service at the University of Vermont Medical Center with a suspected foreign body of the right ear. However, physical examination revealed a fibro-osseous lesion filling the middle ear space and a 50-dB conductive hearing loss. A mastoidectomy and middle ear exploration demonstrated destruction of the external auditory canal, exposure of the TMJ capsule, ablation of the hypo- and protympanum by tumor, and extension into the infratemporal fossa. The extent of resection of the lesion resulted in complete exposure of the carotid artery from the infratemporal fossa to the eustachian tube. A canaloplasty, tympanoplasty, meatoplasty, and split-thickness skin grafting were performed. Final pathology was consistent with fibrous dysplasia and postoperative audiometry demonstrated a flat 20-dB conductive hearing loss. The patient was followed until age 8 years, at which time he had no evidence of recurrent fibrous dysplasia. Over the ensuing 4 years, the patient received care through a regional managed health care system and was lost to follow-up. At age 12, the patient re-presented with a 6-week history of complete facial nerve paralysis. An audiogram at this time revealed profound sensorineural hearing loss and complete loss of the ipsilateral compound motor action potential on ENoG

(Fig. 11). A CT scan showed fibrous dysplasia involving the cochlea, petrous apex, and geniculate ganglion (Fig. 12). However, the external auditory canal remained patent and free of disease. The facial nerve was decompressed from the geniculate ganglion to the lateral internal auditory canal and all gross dysplastic bone resected from the petrous apex via a middle fossa approach. One year postoperatively, the patient demonstrated return of facial nerve function to a House-Brackmann grade III level and stabilization of his disease process on serial CT scans.

DISCUSSION

Fibrous dysplasia is a rare lesion of the temporal bone. Review of previously published cases and the 10 new cases demonstrated the following clinical features. The overall average age at presentation was 19.1 years (range, 9 mo–56 yr). Although historical review revealed a slight male predominance (60.4%), there was a female predominance (80%) in the 10 new cases. Hearing loss was the most common presenting complaint, followed in order by a mass in the temporal bone, unilateral otorrhea, and trismus. Two patients with the polyostotic form developed trismus secondary to invasion of the temporomandibular joint and external auditory canal. Stenosis of the external auditory canal was the most common finding, occurring in at least 80% of patients in both the historical and new groups. The 40% rate of canal cholesteatoma and potential for middle ear extension and hearing loss underscores the need for a high index of suspicion in such patients presenting with progressive canal stenosis and conductive hearing loss.



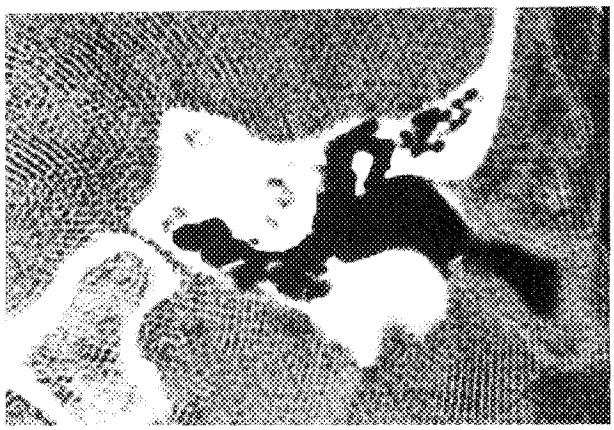


Figure 12. Composite coronal views of right (*top*) and left (*bottom*) bony labyrinth (case 10) in patient with sensorineural hearing loss and acute facial paralysis secondary to monostotic fibrous dysplasia of the right temporal bone. Note encroachment and disruption of normal bony architecture in the region of the cochlea and geniculate ganglion by fibrous dysplasia on the right.

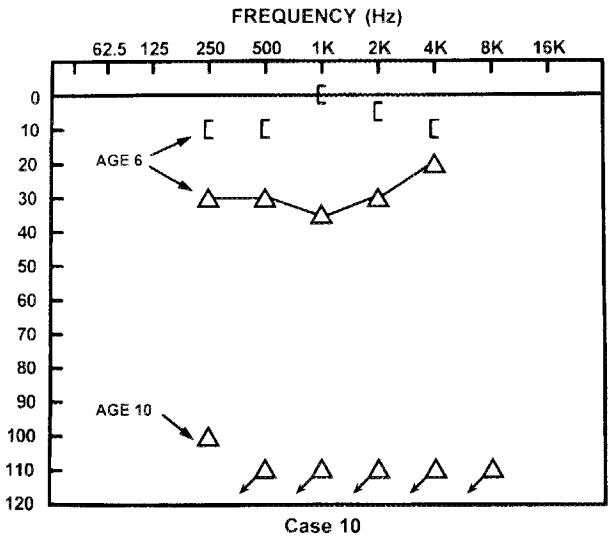


Figure 11. Composite audiogram (case 10) in child with monostotic fibrous dysplasia of the right temporal bone who demonstrated a 20–30 dB air–bone gap after mastoid tympanoplasty at age 6 for treatment of dysplasia–induced otitis media and external auditory canal stenosis. At age 10, the patient re-presented with complete sensorineural hearing loss and acute facial paralysis.

Conductive hearing loss was more common than sensorineural loss. However, 13.9% of the historical cases (6 patients) demonstrated a profound sensorineural hearing loss. Three patients in the new series (cases 2, 9, 10) also demonstrated profound sensorineural hearing loss after initially presenting with a pure conductive loss. The three new patients and the six previously reported cases with sensorineural hearing loss all were found to have radiographic evidence of extensive fibrous dysplasia involving the petrous as well as the mastoid and tympanic portions of the temporal bone. The sensorineural loss was attributable to involvement of the otic capsule or the internal auditory canal.

Indications for surgical intervention in patients diagnosed with fibrous dysplasia of the temporal bone include the presence of conductive hearing loss and cholesteatoma secondary to disease-induced external auditory canal stenosis. The high rate (nearly 90%) of re-stenosis of the external auditory canal

following limited canaloplasty suggests that a more extensive resection is necessary to maintain canal patency and adequate hearing. Of those cases treated via a postauricular mastoidectomy (usually canal wall down) and wide bony canaloplasty with or without split thickness skin grafting, the external canal remained patent in 100% with a follow-up period of 6-months to 10 years. This approach was utilized in case 4 in the present series and has resulted in a patent, trouble free canal 9 months postoperatively despite three previous failed canaloplasty procedures. Such an approach to the maintanence of external auditory canal patency does not preclude future disease progression medially toward the petrous bone with resultant sensorineural hearing loss. This dilemma is exemplified in case 10 where cochlear destruction and profound deafness occurred in a delayed manner despite the early eradication of an apparent lateral temporal bone focus of fibrous dysplasia.

Facial paralysis is unusual even with encroachment on the facial canal by fibrous dysplasia, as exemplified in case 9. However, complete and permanent paralysis has been reported in association with this disease.[29,49] The two such representative historical cases, comprising 4.6% of the total, occurred in association with complete sensorineural hearing loss and extensive petrous apex disease. When all patients are considered, the incidence of facial paralysis or paresis in cases of fibrous dysplasia of the temporal bone was 9.4%. The only reported case of facial nerve decompression following acute facial paralysis in a patient with fibrous dysplasia of the temporal bone (case 10) resulted in a favorable surgical outcome (House-Brackmann grade III) up to 1 year postoperatively. The lack of clinical or radiographic change in this case over a 10-year period, with accelerated activity at puberty, raises interesting speculation concerning hormonal influence.

The potential for delayed geniculate ganglion compression and progression of conductive hearing loss to profound sensorineural hearing loss, often times at or around the time of puberty, mandates long-term follow-up in these patients. Although conductive hearing loss and canal stenosis secondary to fibrous dysplasia, initially limited to the lateral temporal bone, can be successfully ameliorated via postauricular canal wall down mastoidectomy, canaloplasty, and split-thickness skin grafting, long-term disease surveillance is indicated. This should include regular documentation of canal patency, hearing acuity, and cranial nerve integrity. In addition, serial CT scanning is helpful in monitoring the activity of disease and the potential for petrous bone and otic capsule involvement. In cases demonstrating signs or symptoms of early facial nerve compression, electroneurography can be used to document nerve injury and guide the surgeon with regard to the need for facial nerve decompression.

CONCLUSION

Fibrous dysplasia of the temporal bone usually affects patients in the first or second decade of life. The most common findings include stenosis of the bony ear canal in approximately 85%, conductive hearing loss in 80%, and canal cholesteatoma in 40% of cases. Sensorineural hearing loss was found in nearly 17% of cases. A pure conductive hearing loss may be transformed into profound sensorineural hearing loss as the disease progresses to involve the bony labyrinth or internal auditory canal, as documented in three of the newly presented cases. Facial paralysis or paresis complicated 9.4% of cases of fibrous dysplasia of the temporal bone. Surgical treatment of canal stenosis should consist of postauricular canal wall down mastoidectomy, canaloplasty, and split-thickness skin grafting to ensure canal patency. Long-term follow-up of all patients with regard to hearing stability and facial nerve integrity is indicated.

REFERENCES

1. Lichtenstein L: Polyostotic fibrous dysplasia. Arch Surg 1938; 36:874–898.
2. Robbins SL, Cotran RS, Kumar V: Pathologic basis of disease. 3rd Ed. Philadelphia: WB Saunders, 1984.
3. Reitzik M, Lownie JF: Familial polyostotic fibrous dysplasia. J Oral Surg 1975; 40:769–774.
4. Windolz F: Cranial manifestations of fibrous dysplasia of bone. Am J Roentgenol 1947; 58:51–63.
5. Albright F, Butler AM, Hampton AO, Smith P: Syndrome characterized by osteitis fibrosa disseminata, areas of pigmentation, and endocrine dysfunction, with precocious puberty in females. N Engl J Med 1937; 216:727–746.
6. Danon M, Robboy SJ, Kim S, et al: Cushing syndrome, sexual precocity, and polyostotic fibrous dysplasia (Albright syndrome) in infancy. J Pediatr 1975; 87:922–927.
7. Lichtenstein L, Jaffe HL: Fibrous dysplasia of bone. Arch Pathol 1942; 33:777–816.
8. Murray RC, Kirkpatrick HJR, Forrai E: Case of Albright's syndrome (osteitis fibrosa disseminata). Br J Surg 1946; 34:48–57.
9. Changus GW: Osteoblastic hyperplasia of bone. A histochemical appraisal of fibrous dysplasia of bone. Cancer 1957; 10:1157–1161.
10. Schlumberger HG: Fibrous dysplasia of single bones (monostotic fibrous dysplasia). Milit Surgeon 1946; 99:504–527.
11. Lee PA, Van Dop C, Migeon CJ: McCune-Albright syndrome: long-term follow-up. JAMA 1986; 256:2980–2984.
12. Schuknecht HF: Pathology of the ear. 2nd Ed. Philadelphia: Lea & Febiger, 1993:392–396.
13. Fries JW: The roentgen features of fibrous dysplasia of the skull and facial bones: a critical analysis of thirty-nine pathologically proven cases. Am J Roentgenol 1957; 77:71–88.
14. Booth JB: Medical management of sensorineural hearing loss. Part II: Musculo-skeletal system. J Laryngol Otol 1982; 96:773–795.
15. Zappia JJ, LaRouere MJ, Telian SA: Massive ossifying fibroma of the temporal bone. Otolaryngol Head Neck Surg 1990; 103:480–483.

16. Schwartz DT, Alpert M: The malignant transformation of fibrous dysplasia. Am J Med Sci 1964; 247:35–54.
17. Gross CW, Montgomery WW: Fibrous dysplasia and malignant degeneration. Arch Otolaryngol 1967; 85:653–657.
18. Brunner H: Fibrous dysplasia of facial bones and paranasal sinuses. Arch Otolaryngol 1952; 55:43–54.
19. Barrionuevo CE, Marcallo FA, Coelho A, et al: Fibrous dysplasia and the temporal bone. Arch Otolaryngol 1980; 106:298–301.
20. Towson CE: Monostotic fibrous dysplasia of the mastoid and the temporal bone. Arch Otolaryngol 1950; 52:709–724.
21. Skolnik EM, Perrelli SL, Pornoy RA: Fibrous dysplasia of the skull and temporal bone. Ear Nose Throat J 1958; 37:755–759.
22. Kearney HL: Fibrous dysplasia of the temporal bone. Laryngoscope 1959; 69:571–574.
23. Sussman HB: Monostotic fibrous dysplasia of the temporal bone: report of a case. Laryngoscope 1961; 71:68–77.
24. Wong A, Vaughan CW, Strong MS: Fibrous dysplasia of temporal bone. Arch Otolaryngol 1965; 81:131–133.
25. Fluur E, Soderberg G: Monostotic fibrous dysplasia of the mastoid. J Laryngol Otol 1966; 80:678–685.
26. Basek M: Fibrous dysplasia of the middle ear: a case report. Arch Otolaryngol 1967; 85:26–29.
27. Samy LL, Girgis IH, Wasef SA: Fibrous dysplasia in relation to the paranasal sinuses and the ear. J Laryngol Otol 1967; 81:1357–1371.
28. Shiffman F, Aengst FE: Fibrous dysplasia of temporal bone. Arch Otolaryngol 1967; 86:528–534.
29. Cohen A, Rosenwasser H: Fibrous dysplasia of the temporal bone. Arch Otolaryngol 1969; 89:447–459.
30. Tembe D: Fibro-osseous dysplasia of temporal bone. J Laryngol Otol 1970; 84:107–114.
31. Sharp M: Monostotic fibrous dysplasia of the temporal bone. J Laryngol Otol 1970; 84:697–708.
32. Stecker RH: Ossifying fibroma of the middle ear: report of a case. Arch Otolaryngol 1971; 94:80–82.
33. Chatterji P: Massive fibrous dysplasia of the temporal bone. J Laryngol Otol 1974; 88:179–183.
34. Talbot IC, Keith DA, Lord IJ: Fibrous dysplasia of the craniofacial bones: a clinicopathological survey of seven cases. J Laryngol Otol 1974; 88:429–443.
35. Williams DML, Thomas RSA: Fibrous dysplasia. J Laryngol Otol 1975; 89:359–374.
36. Ward PH, Foster C: Monostotic fibrous dysplasia of the temporal bone. Trans Pac Coast Oto-Ophthalmol Soc 1975; 56:147–155.
37. Levine PA, Wiggins R, Archibald RWR, Britt R: Ossifying fibroma of the head and neck: involvement of the temporal bone—an unusual and challenging site. Laryngoscope 1981; 91:720–725.
38. Nager GT, Kennedy DW, Kopstein E: Fibrous dysplasia: a review of the disease and its manifestation in the temporal bone. Ann Otol Rhinol Laryngol [Suppl] 1982; 91(92):1–52.
39. Schrimpf R, Karmody CS, Chasin WD, Carter B: Sclerosing lesions of the temporal bone. Laryngoscope 1982; 92: 1116–1119.
40. Nager GT, Holliday MJ: Fibrous dysplasia of the temporal bone: update with case reports. Ann Otol Rhinol Laryngol 1984; 93:630–633.
41. Lambert PR, Brackmann DE: Fibrous dysplasia of the temporal bone: the use of computerized tomography. Otolaryngol Head Neck Surg 1984; 92:461–467.
42. Wolfson RJ, Berson D, Goodman RS: Temporal bone involvement in polyostotic fibrous dysplasia. Trans Penn Acad Ophthalmol Otolaryngol 1984; 37:60–64.
43. Sataloff RT, Graham MD, Roberts BR: Middle ear surgery in fibrous dysplasia of the temporal bone. Am J Otol 1985; 6:153–156.
44. Smouha EE, Edelstein DR, Parisier SC: Fibrous dysplasia involving the temporal bone: report of three new cases. Am J Otol 1987; 8:103–107.
45. Younus M, Haleem A: Monostotic fibrous dysplasia of the temporal bone. J Laryngol Otol 1987; 101:1070–1074.
46. Pouwels ABPM, Cremers CWRJ: Fibrous dysplasia of the temporal bone. J Laryngol Otol 1988; 102:171–172.
47. Talmi YP, Finkelstein Y, Bar-Ziv J, et al: Monostotic fibrous dysplasia of the temporal bone: pathologic quiz. Arch Otolaryngol 1989; 115:1136–1139.
48. Kessler A, Wolf M, Ben-Shoshan J: Fibrous dysplasia of the temporal bone presenting as an osteoma of the external auditory canal. Ear Nose Throat J 1990; 69:197–199.
49. Reddy KTV, Vinayak BC, Jefferis AF, Grieve DV: Imaging case study of the month: fibrous dysplasia of the temporal bone. Ann Otol Rhinol Laryngol 1994; 103:74–76.

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